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### The Enigma of the Fontan circulation

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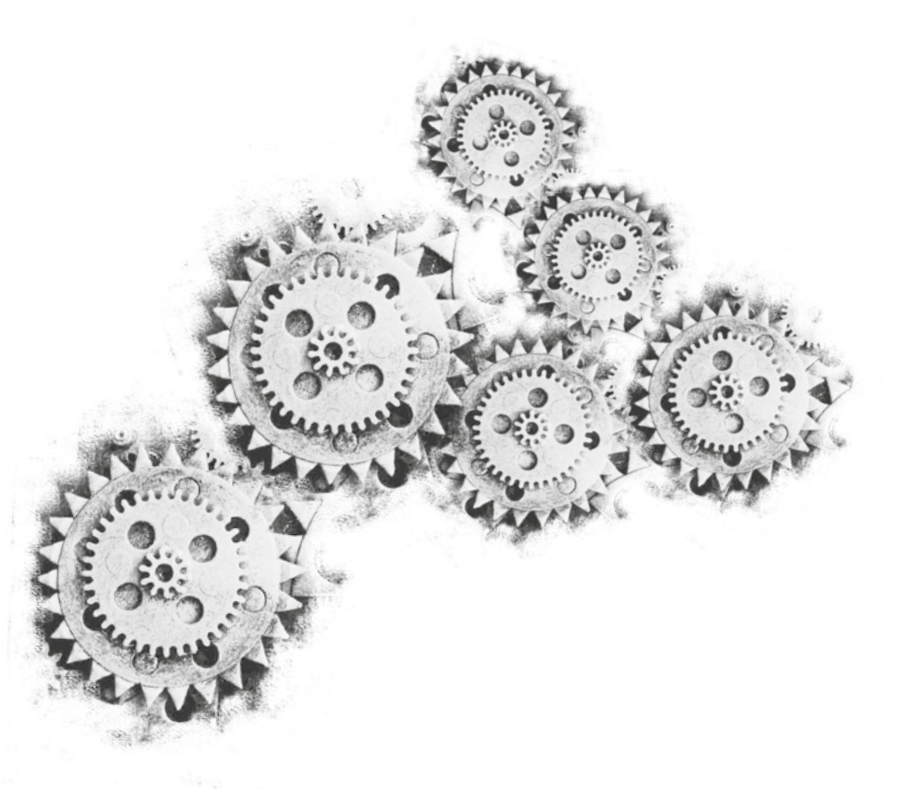
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# 8



## Quality of life and sexual wellbeing in patients with a Fontan circulation

An explorative pilot study



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## Abstract

**O**bjectives: 1) To get an impression of the quality of life and sexual wellbeing in the Fontan population, and 2). To generate hypotheses on the possible pathogenesis of sexual dysfunction for future research.

**Methods:** For this cross-sectional pilot study, questionnaires regarding health-related quality of life, sexual function and fertility/pregnancy were completed by 21 patients with a Fontan circulation > 16 years old, followed at the University Medical Center Groningen, the Netherlands. Semi-structured qualitative interviews were conducted in 8 patients.

**Results:** Adolescents and adults with a Fontan circulation report a health-related Quality of Life and sexual wellbeing generally comparable to normal values. During childhood, most patients experienced physical limitations and the feeling of being an outsider, and frequently faced bullying. Regarding sexual wellbeing, large inter-individual differences were noted in both the questionnaires and interviews. Four interviewed patients (25-30 years old) reported a good sexual wellbeing, whereas the other four interviewed patients (33-47 years old) reported sexual limitations, including erectile dysfunction, low body- and sexual self-esteem and avoidance of sexual intercourse due to arrhythmias.

**Conclusions:** This study generated the following hypotheses for future research: 1). Children and adolescents with a Fontan circulation face more social isolation and bullying than their healthy peers. 2). Fontan patients are fairly well adapted to their physical restrictions and only a minority of these patients experience sexual dysfunction. 3). There is a male/female difference regarding sexual wellbeing in patients with a Fontan circulation. 4). Sexual function in Fontan patients deteriorates with progressive attrition of the Fontan circulation.

## Introduction

Since its invention in 1971, the Fontan circulation has become common practice to treat patients with a univentricular heart who are not suitable for a biventricular repair<sup>1,2</sup>. With the Fontan operation, the systemic venous return is directed to the pulmonary vascular bed without help of a subpulmonary ventricle. The Fontan circulation is characterized by chronically increased central venous pressure and restricted ventricular filling due to the passive pulmonary blood flow, and an increased ventricular afterload, caused by the coupling of the systemic and pulmonary circulation<sup>3</sup>. Various modifications in surgical technique and peri-operative care have been developed over the past decades, and short term outcome has significantly improved<sup>4</sup>. With a growing cohort of patients with a Fontan circulation now reaching adolescence and early adulthood, functional capacity and quality of life has become of increasing interest.

Previous studies showed that patients with a Fontan circulation have an impaired exercise tolerance (around 60% of healthy subjects<sup>5</sup>), and are prone to develop various complications, including arrhythmias<sup>6</sup>. However, quality of life in Fontan patients seems in general well preserved, with 90-100% of the patients within normal limits<sup>7</sup>. One important aspect of quality of life is sexual wellbeing. Previous studies showed that adolescents and young adults with congenital heart diseases have increased concerns regarding fertility, inheritability and pregnancy, may experience a broad range of sexual problems and might lag behind in psychosexual development<sup>8,9</sup>. Although these studies have included a variety of congenital heart disease, no patients with a Fontan circulation were included. This is unfortunate because the unique physiology of the Fontan circulation justifies special attention for their sexual development and wellbeing. Important characteristics in this context include restricted cardiac output and exercise tolerance, the chronic systemic venous congestion and potential autonomic dysregulation in response to the decreased cardiac output<sup>10</sup>. Furthermore, the operations at a young age, frequent hospital visits, and impaired life expectancy might affect patients' development and wellbeing. Finally, menstrual cycle disorders and fertility problems may influence the patients' sexual wellbeing<sup>11</sup>.

In order to fill this gap in our knowledge, we decided to perform an explorative pilot study to investigate sexual function of patients with a Fontan circulation. Its aim was twofold: 1). To get an impression of the sexual wellbeing in a sample of the Fontan population, and 2). To generate hypotheses for future research.

## Method

### Subjects

In 2012-2013, a cross-sectional study was performed among Fontan patients  $\geq 10$  years old who were followed at the outpatient clinics of the University Medical Center Groningen, the Netherlands. Of these consecutive patients, all participants  $\geq 16$  years old were asked to participate in the current explorative pilot study concerning sexual function and wellbeing.

The institutional ethics committee approved this study. It was conducted in accordance with the declaration of Helsinki and written informed consent was obtained from all study participants.

## Procedure

Patient characteristics were collected from medical records and included, gender, date of birth, cardiac anatomic diagnosis, surgical procedures prior to the Fontan completion, type and date of Fontan completion and current medication use.

In order to get an impression of sexual function and wellbeing a multi-method approach was used:

1. General health-related Quality of Life (hrQoL) was assessed using the SF-36 questionnaire. The SF-36 questionnaire includes eight health concepts and for each subdomain, scores ranging from 0 to 100 are calculated. The results of the SF-36 questionnaire were compared to reference values of the Dutch population<sup>12,13</sup>.
2. The Golombok Rust Inventory of Sexual Satisfaction (GRISS) was assessed to measure sexual dissatisfaction and problems in heterosexual women and men<sup>14</sup>. Questions regarding pregnancies, fertility and children were added. After visiting the outpatient clinic, the participating patients were asked to complete the questionnaires at home, allowing for sufficient privacy, and send the completed questionnaires back to the researcher. The results of the GRISS questionnaire of the study population were compared to the reference values of healthy Dutch student couples published by Ter Kuile et al.<sup>15</sup>.
3. In order to generate a clinical impression and hypotheses, ten consecutive patients were asked to participate in semi-structured interviews regarding sexual development and current sexual wellbeing. These interviews were performed by the first author (D.W.). An outline of the interview structure is presented in supplementary file 1.

## Statistical analyses

Patient characteristics were displayed as mean  $\pm$  standard deviation (SD) in normally distributed variables, as median (interquartile range) in skewed variables and as number of patients (percentage of total) in categorical variables. Results of the questionnaire were compared to the reference values using Levene's test to test the equality of variances and t-test analyses to test the equality of the means. A p-value  $< 0.05$  was considered significant. All analyses were performed using SPSS for Windows.

## Results

### Subjects

For the questionnaires, 57 patients were asked to participate, of whom 21 patients were eventually included in the current explorative pilot study. The reasons for exclusion were patient refusal (18 patients), no return of the questionnaire (11 patients) or incomplete filled-in questionnaire (7 patients). The participants were 27 (23-30) years old, with a minimum of 19 years and maximum of 44 years. Further patient characteristics are listed in table 1.

For the interviews, ten randomly selected patients were asked. Two patients refused to participate in the interviews because they would rather not talk about sexual wellbeing. Therefore, the interviews are eventually performed in 8 patients; 3 males and 5 females. The short summaries of the interviews are displayed in the supplementary file 2.

### General wellbeing

The self-reported health related Quality of Life scores of the Fontan patients regarding the domains physical functioning, social functioning, role limitations due to physical problems, role restrictions due to emotional problems, mental health, vitality, pain, and health-change over the last year did not significantly differ from the healthy controls (table 2). The Fontan patients, however, scored significantly lower on general health than their healthy peers ( $p=0.008$ ).

The interviewed patients explained that they currently experience few limitations due to their cardiac condition. They feel that they have a good quality of life in general, despite not being able to work, needing an electronic bicycle or not participating in sports due to their physical restrictions. On the other hand, most of the interviewed patients (6/8) had experienced physical limitations during their childhood and/or puberty. This is illustrated by the following statement by patient G:

*“As a child I realized I was different than the other kids. Now I am used to my limitations, they are a part of who I am and they don’t bother me anymore” (Female, 25)*

and by patient B:

*“I know that my peers have a higher exercise performance than I have, but nowadays, I do not often get confronted with it. This is in contrast to my childhood, when I was confronted with my restrictions on a daily basis.” (Male, 47)*

The physical limitations as a child had a clear consequence for almost all interviewed patients; namely the feeling of being an outsider at primary and/or secondary school. Despite the fact that most patients had (at least some) friends, they were from a young age on aware that they were more restricted during exercise and missed more days at school than their peers.

Consequently, most of the patients (6/8) experienced forms of bullying during either primary or secondary school.

Patient D:

*"I liked going to the primary school, but after two months at the secondary school I did not want to go anymore. I was just not part of the group, and therefore an easy victim. I could not participate in sports activities. The rest of the class biked to the city at lunch time, but I always stayed behind because, at that age, nobody will wait for you lagging behind."* (Female, 31)

**Table 1. Patient characteristics (N=21)**

Male, N(%)	7 (33%)
Female, N(%)	14 (67%)
Diagnosis, N(%)	
TA	9 (43%)
DILV	6 (29%)
AVSD / unbalanced VSD	4 (19%)
PA with IVS	2 (10%)
Ventricular morphology, N(%)	
Left dominant	18 (86%)
Right dominant	3 (14%)
Type of Fontan procedure, N(%)	
TCPC lateral tunnel	13 (62%)
Atriopulmonary connection	5 (24%)
TCPC extracardiac conduit	2 (10%)
Bjork modification	1 (5%)
Age at Fontan procedure, years	4.5 (3.7-6.9)
Current age, years	27 (23-30)
Negative chronotropic arrhythmica, N(%)	7 (33%)

Data expressed as count (% of number), with the number (*n* or *N*) indicated at the top of the column, or as median (25th-75th percentile). APC, atriopulmonary connection; ECC, extracardiac conduit; ED, external diameter; DILV, double-inlet left ventricle; DIRV, double-inlet left ventricle; HLHS, hypoplastic left heart syndrome; LT, lateral tunnel; PA/IVS, pulmonary atresia with intact ventricle septum; TA, tricuspid atresia; TCPC, total cavopulmonary connection.

Despite being bullied, none of the patients felt restrained to make new friends due to their cardiac condition. More importantly, most of the patients did not report any restrains to commit to a friendship or relationship during their adolescence or young adulthood.

### Sexual function and wellbeing

Based on the 21 completed GRISS questionnaires, sexual function and wellbeing in Fontan patients showed no significant differences compared to healthy controls (table 3). However, in the subdomains concerning female dyspareunia and male erectile dysfunction, 2/21 individual patients had a Z-score  $> 2$  SD compared to their healthy peers, causing significantly larger variances than in the healthy controls ( $p=0.019$  respectively  $p=0.008$ ). During the interviews, these large inter-individual differences were noticed as well. Four patients (2 males and 2 females; 25-30 years old) told that they did not experience any restriction or sexual dysfunction. The other patients (1 male and 3 females, 33-47 years old) reported erectile dysfunction (1/7), a very low self-esteem as a consequence of childhood experiences (1/7), and severe arrhythmias affecting sexual desire and perseverance (2/7).

Patients B's concerns about erectile dysfunction:

*“Since my 30th birthday, it is difficult to keep an erection during sexual intercourse. The urologist eventually concluded that it was probably caused by a combination of factors, and one of these was my cardiac condition. Furthermore, the therapeutic options for my problem are very limited due to my cardiac condition. (...) These problems slow me down to take steps in this area with my new partner.” (Male, 47)*

**Table 2. Domains of health-related Quality of Life (N=21)**

	Fontan patients	Z-score Fontan patients	p-value
Physical functioning	73±22	-0.3±0.9	ns
Social functioning	85±16	-0.4±0.8	0.076
Role Physical	76±37	-0.2±1.2	ns
Role Emotional	83±31	-0.2±1.1	ns
Mental Health	78±17	-0.1±1.0	ns
Vitality	65±18	-0.2±0.9	ns
Bodily pain	85±18	0.2±0.6	0.087
General health	59±24	-0.7±1.1	0.008
Health-Change	53±18	0.0±1.1	ns



Regarding the SF-36 subdomains Infrequency, Dissatisfaction, Non-Sensuality, Premature Ejaculation (M), and Anorgasmia (F), no specific problems were reported during the interviews.

In the subdomain Female and Male Avoidance, the scores of the Fontan patients did not show a significant difference with the normal values. However, the two interviewed patients who reported that severe arrhythmias affected their sexual life, indicated that they tend to avoid sexual intercourse. These patients avoided sexual intercourse because they are afraid to instigate or worsen the arrhythmias.

Patient E:

*"Nowadays I tend to avoid sexual intercourse. Every time we have sex, my arrhythmias start again. It really decreases your desire to have sex. I can not enjoy it anymore, because I can not relax, constantly thinking about my cardiac condition." (Female, 46)*

Concerning the SF-36 subdomain Non-Communication, no problems were reported during the interviews about the current relationships of the patients. However, three out of eight interviewed patients reported that they found it difficult to tell about their cardiac condition to a new partner, mostly because they were afraid of their reaction. They thought their new partner would be scared to share a life with someone with a severe cardiac condition, restricted life expectancy, frequent hospital visits and restricted exercise limitations along with sexual impairment (in 1 male patient).

### **Fertility, pregnancy and children**

Three out of the fourteen females who completed the questionnaires had been pregnant. Two of these women sought for help at the fertility clinic. Together, the women had four pregnancies; one ended in a miscarriage (< 20 weeks pregnancy) and the other three babies were born prematurely (at 27, 31 and 35 weeks pregnancy). Of the latter three babies, one died after two months due to meningitis and the other two are still alive. Eight of the female participants reported that they wish to have children in the future, of whom five females were discouraged by their physician to have children due to their heart condition.<sup>16</sup>

Of the five interviewed women, two reported a low self-esteem due to the fact that they could not, or are discouraged to, become pregnant.

Patient Fs' answer to the question if she has to offer enough as a partner:

*"I find that very difficult. My partner and I cannot have any children, because of my cardiac condition. I struggle with the fact that I cannot offer him children, while another woman could. It would have been easier for me if he also had a fertility problem." (Female, 33)*

Based on the questionnaires, two of the males reported that they established a pregnancy

**Table 3. Sexual function and wellbeing**

	Fontan patients (N=21)	Controls (N=68)	p-value
<b>FEMALES</b>			
Avoidance	5.9±2.8	5.5±1.3	0.393
Non sensuality	5.1±1.9	5.5±1.6	0.430
Dissatisfaction	6.6±2.9	6.6±2.2	0.998
Dyspareunia	6.5±3.3	5.2±1.7	0.191
Anorgasmia	9.4±3.7	10.0±3.6	0.559
Non-communication	4.7±2.6	4.5±1.6	0.746
Infrequency	6.1±2.0	5.3±1.7	0.111
<b>MALES</b>			
Avoidance	4.3±0.8	4.8±1.0	0.292
Non sensuality	4.8±1.0	5.1±1.3	0.671
Dissatisfaction	8.2±1.9	8.0±2.7	0.914
Erectile dysfunction	8.2±3.2	5.5±1.4	0.096
Premature Ejaculation	7.8±2.5	7.7±2.2	0.867
Non-communication	3.8±1.3	4.4±1.6	0.441

in a woman, both without any help of fertility improving therapy. In both cases, the child was born healthy, without any heart defect. Three of the other males reported a desire for children in the future. None of males was, because of their heart condition, discouraged by their physician to get children.

During the interviews, none of the males was concerned about fertility or raising children.

## Discussion

In the current study we found Fontan patients to report a health-related Quality of Life and sexual wellbeing generally comparable to normal values. However, during childhood, most patients experienced physical limitations and the feeling of being an outsider.

Regarding sexual wellbeing, large inter-individual differences were noted especially in the subdomains dyspareunia and erectile dysfunction of the questionnaire, and regarding erectile dysfunction, avoidance, and self-esteem during the interviews. Based on the current study, some impressions could be sketched:

Fontan patients face daily restrictions and co-morbidities, associated with the Fontan circulation. Despite these restrictions, the current study confirmed that these patients seem to experience a generally well preserved health-related Quality of Life. However, the interviewed patients pointed out that physical restrictions were more prominent during childhood, where they consequently felt an outsider in their peer-group and were often bullied. This has been previously recognized in other patients with chronic illness during childhood, including children with chronic lung disease<sup>17</sup> and patients with juvenile idiopathic arthritis<sup>18</sup>. In children with chronic illness, their sense of normalcy is related to the degree in which the disease interfered with participation in school or play<sup>18</sup>. Furthermore, the number of health issues in children is related to the chance of peer rejection and bullying<sup>19</sup>, and might be associated with depressive symptoms which have been described in Fontan patients<sup>20</sup>. To our knowledge, the high prevalence of social isolation and bullying of Fontan patients is not previously recognized. These are important issues needing attention because they can influence adult self-esteem and mental health outcomes<sup>21</sup>. In future research, the benefit of shared-decision making and promotion of self-efficacy might be of great interest to help the patients with these issues<sup>22</sup>.

The operation(s) at a young age and the abnormal circulation can potentially affect both psychological and physical aspects of the patients' wellbeing. Psychologically, the impaired life expectancy, body- and self-esteem, performance anxiety and impaired fertility might be important factors. Physically, the increased central venous pressure, limited exercise capacity and medication use might impair sexual function. In general, sexual function and wellbeing seemed well preserved in the current study. Some of the patients experienced a limited exercise capacity during sexual intercourse, but this did not impact their sexual wellbeing. This is in line with results from previous studies on a variety of diseases, including congenital heart disease<sup>23</sup> and metastatic midgut carcinoid tumours<sup>24</sup>.

On the other hand, large inter-individual differences were noted and sexual dysfunction was described by several patients on an individual level. Sexual impairment included erectile dysfunction, low body- and sexual self-esteem and avoidance of sexual intercourse. In patients with other congenital heart diseases, it has been reported that 10% of the males suffer from erectile dysfunction, potentially caused by medication use, impaired cardiac output and restricted functional capacity<sup>25,26</sup>. Additionally, in Fontan patients, erectile dysfunction might be provoked by a dysregulation of the autonomic nervous system, chronic systemic venous congestion and endothelial dysfunction<sup>27,28</sup>. One of the three males who were interviewed was unable to maintain an erection during sexual intercourse. This problem affected his sexual life and he was very limited in the pharmacological therapeutic options (e.g. Sildenafil) for his problem.

In women with congenital heart disease, sexual dysfunction, including dyspnea, arrhythmias, fatigue and syncope, and lower self-esteem are previously recognized<sup>23,29</sup>. In the current study, the females reported predominantly mental components which affected their esteem of their sex life and partnership (patients C and F). Their main concern being their inability to offer children to their partners. High concerns regarding fertility and pregnancy are also reported in patients with other congenital heart defects<sup>30</sup>. However, those patients were more concerned about their own health and their ability to carry the baby to term, whereas women with a Fontan circulation were discouraged to become pregnant and were concerned that their partner might consider having children with another woman.

Interestingly, four interviewed patients reported no sexual dysfunction, suggesting that sexual function can be well preserved in the Fontan circulation. In the Fontan circulation, gradual attrition of the circulation has been suggested, manifesting itself by various circulatory complications, including gastro-enteric complications, arrhythmias, and deteriorating functional capacity. The four patients who did not report sexual dysfunction, were the youngest interviewees and in none of these patients adverse sequelae of the Fontan circulation had developed (yet). Therefore, gradual attrition of the Fontan circulation may be associated with progressive sexual (dys)function of the Fontan patients over time. This is also confirmed by patients C and E, who explained that their arrhythmias and the anxiety for a new onset of arrhythmias severely impact their sexual desire and activity, as well as by patient B, who suffers from erectile dysfunction from the age of thirty.

Finally, it is important to notice that the patients were very well-spoken regarding their experiences as a child or youngster with a congenital heart defect. However, when the interviewer addressed their current status of “what they have made of their lives” or “how they address their problems”, the patients found it more difficult to find words and describe their feelings and functioning. Furthermore, the current study included a sample of 21 patients out of 62 patients  $\geq 16$  years old in our Fontan cohort, causing a potential selection bias. These issues might have caused an underrepresentation of psychosocial or sexual problems. However, the current study was not designed to collect quantitative data, but aimed at generating first impressions and hypotheses regarding sexual wellbeing in Fontan patients. Future studies have to be conducted to investigate the prevalence of sexual problems and to identify whether sexual wellbeing of Fontan patients differs from patients with other congenital heart defects.

Altogether, the results of this explorative pilot study addressing quality of life, sexual wellbeing and fertility/pregnancy in Fontan patients, generated the following hypotheses for future research:

- 1). In particular children and adolescents with a Fontan circulation face more social isolation and bullying than their healthy peers.
- 2). Fontan patients adapt fairly well to their physical restrictions and only a minority of these patients experience sexual dysfunction.

- 3). There is a male/female difference regarding sexual wellbeing in patients with a Fontan circulation. In males, physical limitations predominate and in females psychosocial limitations predominate.
- 4). Sexual function in Fontan patients deteriorates with progressive attrition of the Fontan circulation.

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## Supplementary file 1. Interview structure

### Introduction

What is it like for you to have a cardiac condition?

What does it mean in daily life?

### Childhood/puberty

How would you describe your childhood?

How would you describe your family?

Did your environment stimulate you to discover your world, participate in society and make friends?

How would you describe your life at the elementary school?

Did you have friends? Did you participate in sport activities?

How would you describe your high school experiences?

Did you have friends? Did you participate in sport activities?

How did you feel among peers?

### Sexual development

Did you have girlfriends/boyfriends during your teenage years?

First masturbation experience

First sexual experience with a partner

Do you think your cardiac condition affected your sexual development?

How would you describe your sexual wellbeing during your adolescence?

Do you think your cardiac condition withheld you from making friends and committing to relationships?

### Self-esteem

Do you like yourself as a person? And as a partner?

What do you have to offer as a person? And as a partner?

Do you think your cardiac condition impacts on what you have to offer?

### Current sexual wellbeing

How would you describe your current sexual wellbeing?

Does your cardiac condition affect your sexual life? Which factors have the greatest impact?

Can you enjoy sexual activities?

Do you feel anxious for/during sexual intercourse?

Do you experience bodily pain during sexual intercourse?

Do you experience limitations during sexual intercourse due to restricted exercise tolerance?

Females: Do you experience perineal or vaginal pain during sexual intercourse?

Males: Can you get and hold an erection during sexual intercourse?

Do you feel that the health professionals should pay more attention to psychosocial and sexual issues?



## Supplementary file 2. Interview summaries

### Patient A

This 26-years-old male was interviewed together with his female partner. He did not feel any different than the other children at school or the playground during his childhood and puberty, he was always able to participate in sports and stimulated by his parents to discover the world around him.

His first sexual activities with himself were between 12-14 years of age, and with a partner around 16-17 years of age. He does not have any complaints regarding sexual activities, more specifically, he is not afraid to exert and does not experience (chest) pain during sex. His cardiac condition does not prevent him from committing to a relationship and he feels he can offer as much to his partner and the rest of the world as a healthy individual would.

### Patient B

Patient B is a 47-year-old male, who is divorced from his wife a few years ago and now has a new female partner. Concerning his childhood and puberty, he experienced severe physical limitations, he was wheelchair bound until the age of eight years (when he was operated). Also after the operation he was not able to participate in sports or any outdoor play with the other children. He had more female friends than male friends, and he thinks it may be related to his impossibility to participate in the rough games of the boys.

He masturbated for the first time around 11-12 years old and was 18 years old at the first sexual intercourse with a partner. At that time, no specific complaints regarding sexual intercourse are put forward by the patient. From the age of 30 years, he had a relationship with the female he eventually married and started to notice that it was difficult to keep an erection during sexual intercourse. The urologist concluded that this presumably originated from a combination of high expectations and not-optimal cardiac condition. The patient stressed that exercise tolerance was not a limiting factor.

With his current new partner, he did not have sexual intercourse yet. During the interview he admitted to himself that his concerns (mainly spontaneous issues) hinder him from any progress in that field.

### Patient C

This 36-year-old female was in a long-term relationship with a male. She experienced mainly physical restrictions during childhood/puberty, and was not able to participate in sports activities with her peers. Meanwhile, she indicated that she disregarded her limitations until she was 19 years old, when she faced the first arrhythmias. She was not able to finish her education due to her physical limitations.

Furthermore, the arrhythmias affect her sexual life, in which she feels restrained to exercise because she is afraid the arrhythmias will emerge. She struggles with the fact that she can not offer children to her partner (due to the Amiodaron), and can not participate in sport activities with him. She feels that her frequent and long hospitalization during childhood has formed her as an adult. She indicates that her

fight against the operation/catheterization/venous injections were not listened to, and consequently, she now feels that her opinion does not matter. The only thing she can do is to be grateful to the people surrounding her, and that she does not deserve the little extras in life because she does not do the work to earn the extras.

#### **Patient D**

This female is 30 years old, and has a relationship with a male for the past ten years. At primary school, she was not able to participate in the (sport) activities of her peers, and was transferred to a school for children with special needs, where she fitted in very well. However, at secondary school, she went to a public school again and faced bullying for a long time because she could not keep up with her classmates. She did not have many friends.

The patient thinks that her cardiac conditions did not influence her sexual development and current wellbeing. She can sustain the activity until the end, she is not afraid of pain or cardiac related problems. She feels she can offer her partner and her environment just as much as a healthy person would.

#### **Patient E**

A 46-year-old female who has a male partner for 27 years now. During the interview she revealed that she had a good childhood, despite being bullied because of her physical limitations and cyanosis. She did participate in sport activities at school, but was always picked last for the team selections. She went through a relatively normal puberty, including partying, dancing, alcohol and cigarette smoking.

Her first sexual experience with a partner was at the age of 16 years, and she immediately got pregnant. She delivered a baby girl when she was 17 years old, three months prematurely. The baby died. Later, when she was 20 years old, she wanted to get pregnant again but did not succeed. Eventually, at the age of 30 years, she was pregnant of her son. Her son was born 1 month premature, and survived. During this latest pregnancy she developed arrhythmias, which have restricted her in her daily activities ever since. Currently, the fear for recurrent arrhythmias together with her bad exercise tolerance restrains her sexual activities. As a result, she does not enjoy sex anymore. She feels that she does not have to offer much to her partner, because of her bad exercise tolerance. However, she feels she is emotionally stronger than many of her peers.

#### **Patient F**

33-year-old female. During her years at primary school, she was continuously bullied, had no friends and was mostly alone (or with her little brother) at the play yard. When she was 12 years, she had a minor stroke. Since then, her parents did not let her ride the bike to school but she had to go by bus to her secondary school. At the secondary school, she was still alone quite often, but also developed some friendships.

She thinks that her cardiac condition did not influence her sexual development during puberty. Nowadays, she realizes that her sexual desire is lower than her partners', she blames that on her medication use (Burinex, Spironolacton, Sintrom, Slow-K, Keppra). Furthermore, she experiences perianal pain during

sexual intercourse and recognizes that her restricted exercise tolerance influences their sexual activities, which further decreases her sexual desire. In her role as a partner, she feels sorry that she can not offer children to her husband.

### **Patient G**

This 25-year-old female explains that she felt different than her peers during childhood, she was not part of the group. The other children blamed her for not participating in the sport activities and bullied her. During puberty, she did join her friends to the pub at Friday, but she still felt left out at school. She admits that her family is not very warm and welcoming, because they do not talk much, especially not about her cardiac condition.

Regarding relationship and sex, she is a little hesitant to start new relationships due to the fact that she does not like telling people about her heart surgery, because she is afraid that they will judge her. For her current boyfriend, she feels she is a good partner and had enough to offer both mentally and physically. Concerning pregnancy and children, she was told that there are increased risks but no strong contraindications, as long as she is stable and functioning as good as she is now.

### **Patient H**

Patient H is a 30-year old male, married to a female and father of a 5 month-old son. He has very warm feelings regarding his childhood and parents. His parents have always stimulated their son to participate in the society and have taught him that it is more important to enjoy life than to be afraid of dying. Patient H has always participated at school gym, and was a popular kid with lots of friends, including girlfriends when he entered puberty. He never felt withheld to commit to a relationship, but he did feel a hurdle when he had to explain his cardiac condition to a new (girl)friend. Patient H has never experienced sexual problems, especially not concerning erectile function, perseverance, and anxiety for cardiac problems. Despite the fact that he experiences a good quality of life, he was very afraid that his son would have the same congenital heart disease. He explains he could not bear the idea that he would be the instigator of his son's sickness. He is delighted that his son is healthy.



